

Product datasheet

Recombinant Human PAF-1 protein ab159291

1 Image

Description

Product name	Recombinant Human PAF-1 protein
Expression system	Wheat germ
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	<pre>MASRKENAKSANRVLRLISQLDALELNKALEQLVWSQFTQ CFHGFKPGLLA RFEPEVKACLWVFLWRFTIYSKNATV/GQSVLNKYKNDFS PNLRYQPPSK NQKIWYAVCTIGGRWLEERCYDLFRNHHLASFGKVKQCV NFVIGLLKLG LINFILFQRGKFATLTERLLGIHSVFCKPQNIREVGFYMN RELLWHGF AEFLIFLLPLINVQKLKAKLSSWCIPLTGAPNSDNTLATSGK ECALCGEW PTMPHTIGCEHIFCYFCAKSSFLFDVYFTCPKCGTEVHSLQ PLKSGIEMS EVNAL</pre>
Amino acids	1 to 305
Tags	GST tag N-Terminus

Specifications

Our [Abpromise guarantee](#) covers the use of **ab159291** in the following tested applications. The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Western blot ELISA
Form	Liquid
Additional notes	This product was previously labelled as PEX2.

Preparation and Storage

Stability and Storage

Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.31% Glutathione, 0.79% Tris HCl

General Info

Function

Somewhat implicated in the biogenesis of peroxisomes.

Involvement in disease

Defects in PEX2 are the cause of peroxisome biogenesis disorder complementation group 5 (PBD-CG5) [MIM:170993]; also known as PBD-CGF. PBD refers to a group of peroxisomal disorders arising from a failure of protein import into the peroxisomal membrane or matrix. The PBD group is comprised of four disorders: Zellweger syndrome (ZWS), neonatal adrenoleukodystrophy (NALD), infantile Refsum disease (IRD), and classical rhizomelic chondrodysplasia punctata (RCDP). ZWS, NALD and IRD are distinct from RCDP and constitute a clinical continuum of overlapping phenotypes known as the Zellweger spectrum. The PBD group is genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies.

Defects in PEX2 are a cause of Zellweger syndrome (ZWS) [MIM:214100]. ZWS is a fatal peroxisome biogenesis disorder characterized by dysmorphic facial features, hepatomegaly, ocular abnormalities, renal cysts, hearing impairment, profound psychomotor retardation, severe hypotonia and neonatal seizures. Death occurs within the first year of life.

Defects in PEX2 are a cause of infantile Refsum disease (IRD) [MIM:266510]. IRD is a mild peroxisome biogenesis disorder (PBD). Clinical features include early onset, mental retardation, minor facial dysmorphism, retinopathy, sensorineural hearing deficit, hepatomegaly, osteoporosis, failure to thrive, and hypocholesterolemia. The biochemical abnormalities include accumulation of phytanic acid, very long chain fatty acids (VLCFA), di- and trihydroxycholestanoic acid and pipecolic acid.

Sequence similarities

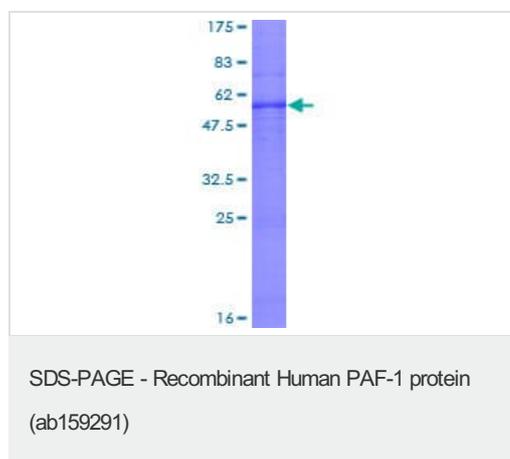
Belongs to the pex2/pex10/pex12 family.

Contains 1 RING-type zinc finger.

Cellular localization

Peroxisome membrane.

Images



ab159291 on a 12.5% SDS-PAGE stained with Coomassie Blue.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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- Response to your inquiry within 24 hours

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